

# Ernesto Bongarzone

## List of Publications by Year in descending order

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57  
papers

2,073  
citations

201674  
27  
h-index

243625  
44  
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58  
all docs

58  
docs citations

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times ranked

2372  
citing authors

#	ARTICLE	IF	CITATIONS
1	Psychosine Accumulates in Membrane Microdomains in the Brain of Krabbe Patients, Disrupting the Raft Architecture. <i>Journal of Neuroscience</i> , 2009, 29, 6068-6077.	3.6	140
2	Recent progress on lipid lateral heterogeneity in plasma membranes: From rafts to submicrometric domains. <i>Progress in Lipid Research</i> , 2016, 62, 1-24.	11.6	134
3	miR-219 Cooperates with miR-338 in Myelination and Promotes Myelin Repair in the CNS. <i>Developmental Cell</i> , 2017, 40, 566-582.e5.	7.0	129
4	Central nervous system myelination in mice with deficient expression of Notch1 receptor. <i>Journal of Neuroscience Research</i> , 2002, 67, 309-320.	2.9	121
5	Neuronal inclusions of $\alpha$ -synuclein contribute to the pathogenesis of Krabbe disease. <i>Journal of Pathology</i> , 2014, 232, 509-521.	4.5	89
6	The Sphingolipid Psychosine Inhibits Fast Axonal Transport in Krabbe Disease by Activation of GSK3 $\alpha$ and Deregulation of Molecular Motors. <i>Journal of Neuroscience</i> , 2013, 33, 10048-10056.	3.6	80
7	Combined hematopoietic and lentiviral gene transfer therapies in newborn Twitcher mice reveal contemporaneous neurodegeneration and demyelination in Krabbe disease. <i>Journal of Neuroscience Research</i> , 2009, 87, 1748-1759.	2.9	72
8	Vitamin C Transporters, Recycling and the Bystander Effect in the Nervous System: SVCT2 versus Gluts. <i>Journal of Stem Cell Research &amp; Therapy</i> , 2014, 04, 209.	0.3	67
9	Psychosine, the cytotoxic sphingolipid that accumulates in globoid cell leukodystrophy, alters membrane architecture. <i>Journal of Lipid Research</i> , 2013, 54, 3303-3311.	4.2	61
10	AAVrh10 Gene Therapy Ameliorates Central and Peripheral Nervous System Disease in Canine Globoid Cell Leukodystrophy (Krabbe Disease). <i>Human Gene Therapy</i> , 2018, 29, 785-801.	2.7	56
11	Persistence of psychosine in brain lipid rafts is a limiting factor in the therapeutic recovery of a mouse model for Krabbe disease. <i>Journal of Neuroscience Research</i> , 2011, 89, 352-364.	2.9	54
12	Extracellular vesicle fibrinogen induces encephalitogenic CD8 $^{+}$ T cells in a mouse model of multiple sclerosis. <i>Proceedings of the National Academy of Sciences of the United States of America</i> , 2019, 116, 10488-10493.	7.1	54
13	Astrocyte Support for Oligodendrocyte Differentiation can be Conveyed via Extracellular Vesicles but Diminishes with Age. <i>Scientific Reports</i> , 2020, 10, 828.	3.3	53
14	Autonomic Denervation of Lymphoid Organs Leads to Epigenetic Immune Atrophy in a Mouse Model of Krabbe Disease. <i>Journal of Neuroscience</i> , 2007, 27, 13730-13738.	3.6	51
15	Long-Term Improvement of Neurological Signs and Metabolic Dysfunction in a Mouse Model of Krabbe's Disease after Global Gene Therapy. <i>Molecular Therapy</i> , 2018, 26, 874-889.	8.2	50
16	Peripheral Neuropathy in the Twitcher Mouse Involves the Activation of Axonal Caspase 3. <i>ASN Neuro</i> , 2011, 3, AN20110019.	2.7	48
17	Sulfatides in extracellular vesicles isolated from plasma of multiple sclerosis patients. <i>Journal of Neuroscience Research</i> , 2016, 94, 1579-1587.	2.9	45
18	Psychosine induces the dephosphorylation of neurofilaments by deregulation of PP1 and PP2A phosphatases. <i>Neurobiology of Disease</i> , 2012, 46, 325-335.	4.4	44

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19	Intrathecal administration of AAV/GALC vectors in 10-11 day-old twitcher mice improves survival and is enhanced by bone marrow transplant. <i>Journal of Neuroscience Research</i> , 2016, 94, 1138-1151.	2.9	42
20	Krabbe disease successfully treated via monotherapy of intrathecal gene therapy. <i>Journal of Clinical Investigation</i> , 2020, 130, 4906-4920.	8.2	41
21	Beyond Krabbe's disease: The potential contribution of galactosylceramidase deficiency to neuronal vulnerability in late-onset synucleinopathies. <i>Journal of Neuroscience Research</i> , 2016, 94, 1328-1332.	2.9	39
22	Macrophages Expressing GALC Improve Peripheral Krabbe Disease by a Mechanism Independent of Cross-Correction. <i>Neuron</i> , 2020, 107, 65-81.e9.	8.1	39
23	Krabbe disease: New hope for an old disease. <i>Neuroscience Letters</i> , 2021, 752, 135841.	2.1	34
24	MMP-3 mediates psychosine-induced globoid cell formation: Implications for leukodystrophy pathology. <i>Glia</i> , 2013, 61, 765-777.	4.9	33
25	Aberrant Production of Tenascin-C in Globoid Cell Leukodystrophy Alters Psychosine-Induced Microglial Functions. <i>Journal of Neuropathology and Experimental Neurology</i> , 2014, 73, 964-974.	1.7	30
26	Mechanism of Neuromuscular Dysfunction in Krabbe Disease. <i>Journal of Neuroscience</i> , 2015, 35, 1606-1616.	3.6	30
27	Characterization and application of a disease-cell model for a neurodegenerative lysosomal disease. <i>Molecular Genetics and Metabolism</i> , 2014, 111, 172-183.	1.1	29
28	Î±-Synuclein interacts directly but reversibly with psychosine: implications for Î±-synucleinopathies. <i>Scientific Reports</i> , 2018, 8, 12462.	3.3	28
29	Psychosine enhances the shedding of membrane microvesicles: Implications in demyelination in Krabbe's disease. <i>PLoS ONE</i> , 2017, 12, e0178103.	2.5	28
30	Inhibition of IGF-1-PI3K-Akt-mTORC2 in lipid rafts increases neuronal vulnerability in a genetic lysosomal glycosphingolipidosis. <i>DMM Disease Models and Mechanisms</i> , 2019, 12, .	2.4	26
31	A microglial hypothesis of globoid cell leukodystrophy pathology. <i>Journal of Neuroscience Research</i> , 2016, 94, 1049-1061.	2.9	24
32	Analysis of age-related changes in psychosine metabolism in the human brain. <i>PLoS ONE</i> , 2018, 13, e0193438.	2.5	24
33	Waning efficacy in a long-term AAV-mediated gene therapy study in the murine model of Krabbe disease. <i>Molecular Therapy</i> , 2021, 29, 1883-1902.	8.2	22
34	Brainstem development requires galactosylceramidase and is critical for pathogenesis in a model of Krabbe disease. <i>Nature Communications</i> , 2020, 11, 5356.	12.8	21
35	SVCT2 Expression and Function in Reactive Astrocytes Is a Common Event in Different Brain Pathologies. <i>Molecular Neurobiology</i> , 2018, 55, 5439-5452.	4.0	20
36	Detection of the Neurotoxin Psychosine in Samples of Peripheral Blood: Application in Diagnostics and Follow-up of Krabbe Disease. <i>Archives of Pathology and Laboratory Medicine</i> , 2012, 136, 709-710.	2.5	19

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37	Hematopoietic Stem cell transplantation and lentiviral vectorâ€based gene therapy for Krabbe's disease: Present convictions and future prospects. <i>Journal of Neuroscience Research</i> , 2016, 94, 1152-1168.	2.9	18
38	Unconventional Neurogenic Niches and Neurogenesis Modulation by Vitamins. <i>Journal of Stem Cell Research &amp; Therapy</i> , 2014, 04, 184.	0.3	17
39	SVCT2 Overexpression in Neuroblastoma Cells Induces Cellular Branching that is Associated with ERK Signaling. <i>Molecular Neurobiology</i> , 2016, 53, 6668-6679.	4.0	15
40	The Role of Vesicle Trafficking and Release in Oligodendrocyte Biology. <i>Neurochemical Research</i> , 2020, 45, 620-629.	3.3	15
41	How membrane dysfunction influences neuronal survival pathways in sphingolipid storage disorders. <i>Journal of Neuroscience Research</i> , 2016, 94, 1042-1048.	2.9	14
42	microRNA-219 Reduces Viral Load and Pathologic Changes in Theiler's Virus-Induced Demyelinating Disease. <i>Molecular Therapy</i> , 2018, 26, 730-743.	8.2	13
43	Standard-flow LC and thermal focusing ESI elucidates altered liver proteins in late stage Niemannâ€Pick, type C1 disease. <i>Bioanalysis</i> , 2019, 11, 1067-1083.	1.5	13
44	Synaptic failure: The achilles tendon of sphingolipidoses. <i>Journal of Neuroscience Research</i> , 2016, 94, 1031-1036.	2.9	12
45	Fluid levity of the cell: Role of membrane lipid architecture in genetic sphingolipidoses. <i>Journal of Neuroscience Research</i> , 2016, 94, 1019-1024.	2.9	11
46	Lead Optimization of Benzoxazolone Carboxamides as Orally Bioavailable and CNS Penetrant Acid Ceramidase Inhibitors. <i>Journal of Medicinal Chemistry</i> , 2020, 63, 3634-3664.	6.4	11
47	Psychosine remodels model lipid membranes at neutral pH. <i>Biochimica Et Biophysica Acta - Biomembranes</i> , 2018, 1860, 2515-2526.	2.6	9
48	Synaptic Function and Dysfunction in Lysosomal Storage Diseases. <i>Frontiers in Cellular Neuroscience</i> , 2021, 15, 619777.	3.7	9
49	Mass spectrometry imaging and LC/MS reveal decreased cerebellar phosphoinositides in Niemann-Pick type C1-null mice. <i>Journal of Lipid Research</i> , 2020, 61, 1004-1013.	4.2	7
50	The Pathogenic Sphingolipid Psychosine is Secreted in Extracellular Vesicles in the Brain of a Mouse Model of Krabbe Disease. <i>ASN Neuro</i> , 2022, 14, 175909142210878.	2.7	7
51	Deregulation of signalling in genetic conditions affecting the lysosomal metabolism of cholesterol and galactosyl-sphingolipids. <i>Neurobiology of Disease</i> , 2020, 146, 105142.	4.4	6
52	AAV-Mediated GALC Gene Therapy Rescues Alpha-Synucleinopathy in the Spinal Cord of a Leukodystrophic Lysosomal Storage Disease Mouse Model. <i>Frontiers in Cellular Neuroscience</i> , 2020, 14, 619712.	3.7	5
53	CRISPR-Cas9 Knock-In of T513M and G41S Mutations in the Murine Î²â€Galactosyl-Ceramidase Gene Re-capitulates Early-Onset and Adult-Onset Forms of Krabbe Disease. <i>Frontiers in Molecular Neuroscience</i> , 2022, 15, .	2.9	5
54	Generation of a LacZ reporter transgenic mouse line for the stereological analysis of oligodendrocyte loss in galactosylceramidase deficiency. <i>Journal of Neuroscience Research</i> , 2016, 94, 1520-1530.	2.9	4

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55	An <em>In Vitro</em> Model for the Study of Cellular Pathophysiology in Globoid Cell Leukodystrophy. Journal of Visualized Experiments, 2014, , e51903.	0.3	2
56	Biochemical Analysis of Lipid Rafts to Study Pathogenic Mechanisms of Neural Diseases. Methods in Molecular Biology, 2021, 2187, 37-46.	0.9	2
57	A tribute to the work and life of Dr. Knud H. Krabbe: Advances in genetics, neuropathogenesis, therapies, and clinical management of Krabbe's disease. Journal of Neuroscience Research, 2016, 94, 963-964.	2.9	1